C-EOS

Classification for Early-Onset Scoliosis

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Development and Initial Validation of a Novel Classification System for Early Onset Scoliosis


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Disclosures

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Disclosure: I DO have a financial relationship with a commercial interest.

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**Travel Expenses:** CWSDSG, FoxPSDSG

**Other:** CWSDSG - BOD

POSNA BOD
Improving the Evidence Base in EOS

Development of a Research Infrastructure
Via five parallel efforts

- **Endpoints**: Development/Validation of a Disease-Specific QoL Measure -- EOSQ
- **Equipoise**: Identifying Clinical Equipoise in the Field of EOS
- **Classification-EOS**: Development / Validation of Classification for EOS
- **Complications Classification**: Standardize Way We Define and Report Complications
- **Clinical Trials**: Proximal Anchors: Rib Vs Spine – Retrospective (Prospective Underway)
Purpose of the Classification for EOS (C-EOS)

To classify EOS patients in order to:

1) **Predict** the disease course of individual patients

2) **Prognosticate** and determine beneficiaries of differing treatment modalities

3) **Improve communication** among EOS providers and facilitate research
Key ‘Philosophical’ Aspects of the (C-EOS)

- **Comprehensive**
  Applicable to all EOS pts

- **Practical**
  Utilized in daily practice

- **Prognostic**
  Predictive of course

- **Guide**
  Informs treatment decisions

An EOS ‘One Liner’
Methods: Validation Pathway

Phase 1
Development phase
- Classification proposal
- Pilot agreement studies

Nominal Group Technique: Iterative Surveying and Group Discussion

Phase 2
Reliability and accuracy in clinical setting
- Multicentre agreement study

Reliability Testing

Phase 3
Association with patient outcome(s)
- Clinical studies

Future Work

Iterative Survey & Group Discussion

- **Group Discussion**
  - Proposing Variables
    - POSNA – May 2011

- **Iterative Survey**
  - Assessing Variables
    - May-July 2011

- **Group Discussion**
  - Finalizing Variables
    - ICEOS – November 2011

Iterative input by 24 surgeons
## Results of Variable Identification Survey

<table>
<thead>
<tr>
<th>Variable</th>
<th>Not Useful</th>
<th>Useful</th>
<th>Essential</th>
<th>CVR</th>
<th>Sum of Ranks</th>
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<td>1</td>
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<tr>
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<td>2</td>
<td>-0.73</td>
<td>12</td>
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<tr>
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<td>5</td>
<td>0</td>
<td>-1.00</td>
<td>5</td>
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<tr>
<td>BONE QUALITY</td>
<td>11</td>
<td>4</td>
<td>0</td>
<td>-1.00</td>
<td>4</td>
</tr>
</tbody>
</table>
C-EOS Variables: Etiology

- Challenging variable due to heterogeneous population
- Numerous iterations based on study group feedback

Etiology

- Idiopathic
- HTNM
- LTNM
- Syndromic
- Congenital
C-EOS Variables: Etiology

Congenital/Structural: Curves developing due to a structural abnormality/asymmetry of the spine and/or thoracic cavity (i.e. hemivertebrae, fused ribs, post-thoracotomy, or CDH)

Neuromuscular: Patient with neuromuscular disease (i.e. SMA, Cerebral Palsy, muscular dystrophies, etc.)

Syndromic: Syndromes with known or possible association with scoliosis (including spinal dysraphism)

Idiopathic: No clear causal agent (can include children with a significant co-morbidity that has no defined association with scoliosis)
Cobb Angle: Measurement of major spinal curve in position of most gravity.
C-EOS Variables: Kyphosis

Maximum Total Kyphosis: Between any two levels throughout spine

- (-) <20°
- N: 21-50°
- (+): >50°
C-EOS Variables:
Progression Modifier (Optional)

- **P0:** $< 10^\circ$/yr
- **P1:** 10$^\circ$-$20^\circ$/yr
- **P2:** $>20^\circ$/yr

Minimum of 6 months x-ray follow-up

$$\frac{[\text{Cobb at } t_2] - [\text{Cobb at } t_1]}{[\text{Months between } t_1 \text{ and } t_2]} \times 12 \text{ months/year}$$
**Etiology (In order of priority):**
- **Congenital/Structural:** Curves developing due to a structural abnormality/asymmetry of the spine and/or thoracic cavity; includes hemivertebrae, fused ribs, post-thoracotomy, or CDH.
- **Neuromuscular:** Pts with neuromuscular disease
- **Syndromic:** Syndromes with known or possible association with scoliosis (including spinal dysraphism)
- **Idiopathic:** No clear causal agent (can include children with a significant co-morbidity that has no defined association with scoliosis)

**Cobb Angle (Major Curve):**
1: \(<20^\circ\)
2: 21-50°
3: 51-90°
4: \(\geq 90^\circ\)

**Maximum Total Kyphosis:**
- \((-\): <20°
- \((-\): >20°
- \(+\): >50°

**Progression Modifier (optional):**
- P0: \(<10^\circ/yr\)
- P1: 10-20°/yr
- P2: >20°/yr

**Cobb Angle:** Measurement of major spinal curve in position of most gravity

**Maximum Total Kyphosis:** between any 2 levels

**Annual Progression Ratio Modifier (optional):**

Progression per year;
min. 6 months between observation

\[
\frac{(Cobb @ t_2) - (Cobb @ t_1)}{[t_2-t_1]} \times 12 \text{ months}
\]
CASE 1

History:
- 19 mo old female
- 38wk, C-section
- L thoracotomy for PDA repair @ 4 mo, scoliosis noted post-op
- Acquired rib fusion b/w concave 4th-5th rib

Physical:
- Hypotonic UE and trunk, hypertonic LE
- Rigid right thoracic curve

C7-T6 = 24º

9 months later

Post-PDA surgery
CASE 1: 9 months later

1. Etiology
   • Acquired chest wall deformity → Congenital/structural

2. Cobb Angle
   • 42º → 2

3. Kyphosis
   • Lateral x-ray reveals 35º maximum total kyphosis → N

4. Progression Modifier (optional)
   • \[\frac{(42^\circ - 24^\circ)}{9 \text{ mo.}}\] x 12 = 24º/yr → P2

C/2/N/P2
CASE 2

History
• 4 y/o girl w/ Congenital Myotonic Dystrophy
  • Mother as well

Physical
• Hyperkyphosis
• Bilateral equinus s/p percutaneous heel lengthening
  – 30° of dorsiflexion
• Full ROM at knees and hips
CASE 2

C7-L4 = 50°

1. Etiology
   • Congenital? Neuromuscular?
     Syndromic?

   • Cobb Angle
     • 50° → 2

1. Kyphosis
   • 96° → +

2. Progression Modifier (optional)
   • Not available

N/2/+
### CASE 3

<table>
<thead>
<tr>
<th>History</th>
<th>Physical</th>
</tr>
</thead>
<tbody>
<tr>
<td>• 4 y/o girl w/ Pena-Shokeir Syndrome</td>
<td>• Lays comfortably on table</td>
</tr>
<tr>
<td>• Developmentally delayed</td>
<td>• Stiff left thoracolumbar curvature</td>
</tr>
<tr>
<td>• Right hip dislocation</td>
<td>• Rib cage rests on pelvis</td>
</tr>
<tr>
<td>• Nonambulatory</td>
<td>• Multiple contractures</td>
</tr>
<tr>
<td>• Wheelchair</td>
<td></td>
</tr>
</tbody>
</table>
CASE 3

T9-L4 = 88°

7 months later

Coronal Cobb = 97°

Kyphosis = 26°

Etiology
- Congenital/Structural
- Neuromuscular
- Syndromic
- Idiopathic

Cobb Angle (Major Curve)
1: <20°
2: 21-50°
3: 51-90°
4: >90°

Maximum Total Kyphosis
- N: 21-50°
- (+): >50°

Progression Modifier (optional)
P0: <10°/yr
P1: 10°-20°/yr
P2: >20°/yr
1. Etiology
   • Pena-Shokier Syndrome
     *Syndromic*

2. Cobb Angle
   • $97^\circ \rightarrow 4$

3. Kyphosis
   • $26^\circ \rightarrow N$

4. Progression Modifier (optional)
   • $[\frac{(97^\circ - 88^\circ)}{(7 \text{ mo.})}] \times 12 = 15.4^\circ/\text{yr} \rightarrow P1$

**S/4/N/P1**
Methods: Validation Pathway

Phase 1
Development phase
- Interviews, Literature Review, and Working Session
  - Classification proposal
  - Pilot agreement studies
  - Nominal Group Technique: Iterative Surveying and Group Discussion

Phase 2
Reliability and accuracy in clinical setting
- Multicentre agreement study

Phase 3
Association with patient outcome(s)
- Clinical studies
- Future Work

Reliability Testing

**Purpose:** To assess C-EOS’ ability to prognosticate outcomes in a clinical setting

**Hypothesis:** Timing to VEPTR fixation failure will differ among C-EOS classes
Methods

Design:
- Retrospective review of prospectively enrolled patients
  - Sourced from a national registry, Chest Wall Spinal Deformity Study Group (CWSDSG)

Participants: Enrollees of the CWSDSG from 2005-2011
- Inclusion
  - EOS diagnosis
  - >2 yrs follow-up
  - VEPTR surgery patients
  - Experienced VEPTR proximal fixation failure
Endpoints:

- Time (months) to VEPTR proximal fixation failure
  
  • *Definition*: Radiographic diagnosis of failure by an EOS surgeon requiring operative revision of the rib cradle

Inclusion:

- Of 446 VEPTR patients with adequate follow up,
- 105 with proximal fixation failure

Statistical Analysis:

- Analysis of Variance (ANOVA) for solitary C-EOS variables
- Kaplan-Meier Survivorship Analysis by C-EOS classes w n>3
Neuromuscular Pts Exhibit Rapid Failure

ANOVA
- NM vs. Idiopathic  
  \( p = 0.026 \)
- NM vs. Congenital  
  \( p < 0.001 \)
C-EOS Stratifies Low, Medium, and High Risk

**Time to VEPTR Anchor Failure**

Classes with n>3 variables:
- C3-
- C3N
- C3+
- N3+
- N3N
- N4N
- N4+

**Time to Failure in Months**

**Survival**
Medium Risk of Failure by C-EOS

Medium Risk:
- Congenital/51-90° / Norm & Hyper-kyphosis
  - C3N, C3+
- Neuromuscular / 51-90° / hyperkyphosis
  - N3+
High Risk of Failure by C-EOS

High Risk:
- Neuromuscular / Curve 51-90° & >90° / Norm & hyperkyphosis
  - N3N, N4N, N4+
Conclusions

1. C-EOS able to stratify risk of rapid VEPTR anchor failure
   • Supports validity of C-EOS instrument
   • Potential for use in clinical setting

2. Neuromuscular etiology and curves > 90° as individual variables at high risk of rapid anchor failure

3. With further study, C-EOS may guide treatment decisions and inform providers
C-EOS applied to min. 5 Yr follow up pts:

• **Purpose:** Apply C-EOS to identify trends

• **Methods:**
  – Retrospective review of CWSDSG & GSSG database
  – Min 5 year follow-up

• **Endpoints:**
  – Complications
  – Change in coronal and sagittal curve over time

• **Status:** Pending data collection from CWSDSG and GSSG Registry
Thank You

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